Case 1: Periorbital swelling

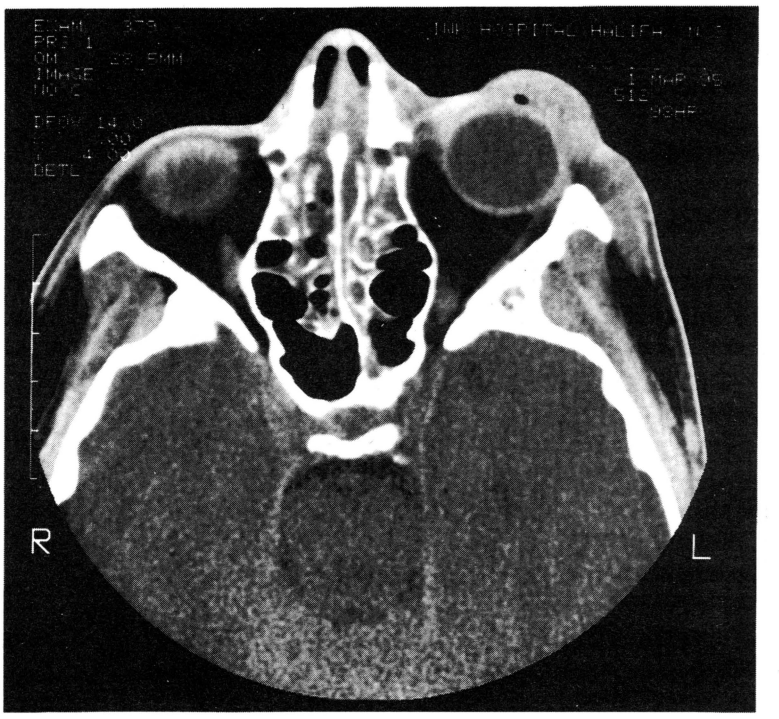
A 13 year old female was admitted with a one day history of left eye swelling. She had an upper respiratory tract infection for the week prior to admission, and developed left eye pain worse with eye movement two days prior to admission. She had been seen in the Emergency Room on the day prior, and was discharged home with a prescription for Ceclor. One month prior to admission she had streptococcal pharyngitis. There was no history of trauma.

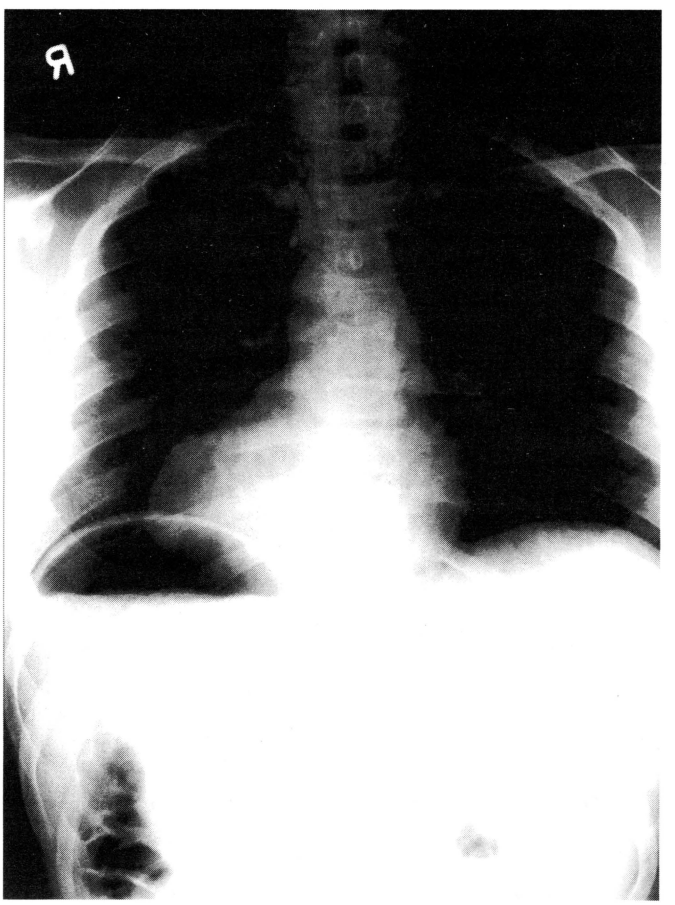
On admission, her oral temperature was 38°C, and the left eyelids were swollen and erythematous. The cornea was clear although there was mild conjunctival edema. Fundi were normal. There were bilateral cervical lymph nodes palpable, but head and neck exam was otherwise normal.

Computed tomogram (CT) of the region is shown at right.

What is the diagnosis? What is appropriate management of this problem?

(Answer on page 51)





Case 2: An abnormal chest x-ray

A 15 year old male was referred to the Otolaryngology Department at the IWK Children's Hospital for assessment of frequent upper respiratory tract infections. He had multiple episodes of sinusitis and pneumonia over the previous 3 years, some requiring intravenous antibiotics. The referring physician had noted an abnormality on the chest radiograph. He had been well earlier in his childhood, and his family history was negative.

The patient was taken to the operating room for bronchoscopy and biopsy, with no unusual findings noted. Routine postoperative chest radiograph is shown.

What does the chest x-ray at left show? Was bronchoscopy and biopsy appropriate for this child? Can this occur spontaneously?

(Answer on page 52)

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Case 2.

Answer: Situs inversus with pneumomediastinum.

The referring physician noticed that the chest x-ray showed situs inversus, as the heart, stomach bubble, and liver are reversed to their normal location. This post-biopsy chest x-ray shows pneumomediastinum as well as situs inversus. A thin dark line representing air is clearest at the right heart border. This patient also had clinical signs of pneumomediastinum including subcutaneous emphysema at the neck and Hamman's sign (a series of precordial crackles synchronous with the heartbeat; 1). This is a known complication of bronchial biopsy, and was treated conservatively with complete resolution in this case.

The bronchial biopsy was performed in order to obtain a sample of respiratory mucosa for electron microscopy to analyse ciliary morphology. This is appropriate investigation for recurrent/chronic respiratory disease, following investigation to rule out allergic disease, cystic fibrosis, and hypogammaglobulinemia as underlying entities (2). The putative diagnosis was Kartagener's syndrome, a combination of situs inversus and primary ciliary dyskinesia, which manifests as chronic sinusitis, purulent rhinitis, otitis media, bronchitis, recurrent pneumonia, bronchiectasis, nasal polyps, and male sterility (2). Two other syndromes are relevant in this clinical scenario; the first is primary ciliary dyskinesia syndrome, which combines abnormal cilia with normal visceral status, and Young syndrome where the clinical picture consists of sinusitis, bronchiectasis, and obstructive azospermia but ciliary ultrastructure is normal (2).

Ciliary beat has a crucial role in the clearance of secretions from sinuses and the lower respiratory tract. The most common abnormality associated with this clinical scenario is absence of dynein arms resulting in ineffective motility although other defects are also possible (2).

Pneumomediastinum can occur secondary to asthma, inhalational drug use, labour, coughing, rapid reduction in atmospheric pressure, or other activities associated with Valsalva maneuvers where terminal alveoli are thought to burst allowing air to track along the pulmonary vasculature into the mediastinum (1,3). Thirty percent of those presenting to the Emergency Room with this condition have no antecedent event, resulting in a diagnosis of spontaneous pneumomediastinum (1). This is most common in young males but has been reported in children as well (3). The most common symptom is stabbing retrosternal pain radiating to the back or shoulders, which is lessened by shallow respiration or leaning forward (3). Dyspnea, dysphagia or a feeling of fullness in the throat may also be present (3,4). Signs include subcutaneous emphysema, Hamman's sign, and decreased cardiac dullness on percussion (3). Diagnosis is made by chest x-ray (1,3,4). Management in the past has consisted of hospitalization for bedrest and analgesia, along with serial chest x-rays (3). A recent review of 17 cases suggests that patients without underlying disease and stable vital signs may be managed with outpatient rest, analgesics, and avoidance of Valsalva producing activities (4). These authors found that serial chest x-rays did not change management in any of their cases, and advise patients to return if symptoms change at which time a chest x-ray is repeated (4).

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Permission to present these cases was given by KD Clarke, Head of the Department of Otolaryngology, Izaak Walton Killam Children's Hospital. Members of the Nova Scotia medical community are invited to submit cases for this diagnostic challenge section.